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Results of trabeculotomy and guarded filtration procedure for glaucoma associated with Sturge-Weber syndrome

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ABSTRACT: Purpose*. To assess the efficacy of trabeculotomy and guarded filtration procedure in the management of glaucoma associated with the Sturge-Weber syndrome.*
Methods. *Six patients (seven eyes) with Sturge-Weber syndrome who underwent glaucoma surgery were retrospectively reviewed. One patient had bilateral glaucoma. Age at the time of operation ranged between 23 days and 9 years. The mean preoperative intraocular pressure was 30.6 mmHg (range 25-38 mmHg). Trabeculotomy and guarded filtration procedure were performed in three eyes each and both techniques were used in one eye.*
Results. *The mean follow-up was 6.3 (range 2-11) years. A single procedure lowered the intraocular pressure and arrested the progression of the disease in five eyes. Two patients required topical antiglaucoma medication. No patient needed reoperation. Two eyes suffered intra- and post-operative temporary choroidal effusions.*
Conclusions. *Trabeculotomy in infancy and guarded filtration procedure in older children effectively stabilized the eyes and prevented further glaucomatous damage in our patients with Sturge-Weber syndrome. (Eur J Ophthalmol 1999, 9: 99-102)*

KEY WORDS: Sturge-Weber syndrome, Glaucoma surgery, Trabeculectomy, Trabeculotomy

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INTRODUCTION

Encephalotrigeminal hemangiomatosis,or Sturge-Weber syndrome (SWS), is one of the most common causes of secondary glaucoma in childhood (l). The syndrome consists of leptomeningeal angiomatosis, nevus flammeus over the cutaneous distribution of the trigeminal nerve and several vascular malformations involving the eye and its adnexae (2). Glaucoma develops in more than 70% of patients with SWS and has a bimodal onset. It usually presents before 2 or after 5 years of age. Interestingly, almost all cases with glaucoma have port-wine stain involving the upper and lower eyelids and no patient with uninvolved eyelids develops glaucoma.(2)

There has been controversy over the best therapeutic approach to control the elevated intraocular pressure and to prevent the ultimate loss of visual function in patients with SWS. Medical management almost invariably fails to control the progression of glaucomatous damage and thus surgical intervention is mandatory. We report our experience in the surgical management of glaucoma in patients with SWS.

METHODS

We retrospectively reviewed the records of all consecutive patients followed between 1979 and 1997 for glaucoma associated with SWS and selected those who required surgery, for further study. The following data were collected for each patient: age at sur-

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gical intervention, visual acuity, intraocular pressures, cup-to-disc ratios, surgery performed, complications of surgery, postoperative follow-up and antiglaucoma medications used.

Surgery was performed when a patient under maximum medical treatment showed rapid enough progression of glaucoma to cause probable deterioration of optic nerve function, documented increasing cupping of the optic nerve head, or when infants with SWS presented with cloudy corneas or enlarged corneal diameters accompanied by elevated intraocular pressure.

Infants were examined under general anesthesia. Intraocular pressures were measured using Schiotz or Perkins tonometers immediately after the induction phase. Older children underwent an office examination which included Snellen visual acuity measurements whenever possible, anterior segment biomicroscopy, direct or indirect ophthalmoscopy, gonioscopy, and applanation tonometry.

In patients with early-onset glaucoma (developing before two years of age), a trabeculotomy as described by Harms was performed (3). Late-onset type glaucoma (developing after five years of age) was addressed by a guarded filtration procedure. In brief, a limbal-based conjunctival flap was prepared and 3X3 mm of scleral flap was dissected. A paracentesis tract was created followed by the excision of approximately 1.5X1.5 mm of clear corneal block. The scleral flap was immediately tied by 3 or 4 sutures depending on the amount of outflow.

Antimetabolite drugs were not used as an adjunct to surgery. Surgical outcome was assessed in terms of postoperative intraocular pressure control, stabilization of the cup-to-disc ratio and visual function and the patient's overall wellbeing.

RESULTS

Seventeen patients with SWS were followed for glaucoma during the period to which this study refers. Six patients (seven eyes) required surgery (Tab. I). All these patients had nevus flammeus on the affected side involving both the upper and lower eyelids. None had choroidal hemangioma. Glaucoma was bilateral in one case (no. 5). The timing of surgery was bimodal. Surgical treatment was inevitable either shortly after birth or after age 5 years, despite a trial of maximum medical therapy. Preoperative intraocular pressures tended to be slightly higher in the late-onset group.

Three eyes underwent trabeculotomy and three eyes had guarded filtration procedure. Combined trabeculotomy-trabeculectomy was done in one patient. Two eyes in the trabeculotomy group (no. 1 and 3) had intraoperative anterior chamber hemorrhages. These hemorrhages were mild and cleared within a week. In the guarded filtration procedure group, one eye (no. 6) had massive intraoperative choroidal effusion. Another (no. 5, right eye) gradually developed choroidal effusion in the early postoperative stage. These two patients had complete resolution of their choroidal effusions after medical treatment.

The mean follow-up was 6.3 (range 2-11) years. No patients required reoperation. Except for two patients, the single procedure in each eye successfully maintained the intraocular pressures so that no further deterioration occurred. In two eyes (no. 2 and 4), the intraocular pressures were lowered closer to traditional "safe" levels with the addition of topical timolol maleate 0.5% and pilocarpine 2% drops. The drugss were begun 6 and 8 months after surgery. There were no late complications. Patients with guarded filtration procedure continued to have functioning blebs during follow-up (Fig. 1). Patients of school age were able to function normally.

DISCUSSION

Secondary glaucomas of childhood including glaucoma caused by SWS are sometimes considered to be the most difficult to manage (1). This is partly because the mechanisms leading to glaucoma in SWS are not fully understood. SWS results from aberrations in the migration and differentiation of neural crest cells (2). According to the widely held "dual" theory, trabeculodysgenesis, indistinguishable from that in primary congenital glaucoma, predominates in early-onset cases, whereas elevated episcleral pressure is the main culprit in late-onset glaucomas (4). Cibis et al (5) found angle abnormalities in infants and observed ultrastructural changes compatible with premature aging of the trabecular meshwork and the Schlemm system in older children, similar to primary open angle glaucoma. Ultrastructural studies on nevus flammeus

TABLE I - SELECTED DATA OF PATIENTS WITH STURGE-WEBER SYNDROME OPERATED FOR GLAUCOMA

Patient	Age at surgery	Laterality	Initial IOP*	Initial VA•	Initial c/d••	Treatment	Additional treatment	Final IOP	Final VA	Final c/d	Follow-up (years)
1	23 days	Left	26	N/A§	0.5	Trabeculotomy	-	15	FF^^	0.2	2
2	3 months	Left	32	FF	N/A	Trabeculotomy	Pilocarpine	15	FF	0.2	2
3	9 months	Left	30	FF	0.6	Trabeculotomy	-	18	20l60	0.7	11
4	6 years	Right	28	20/60	0.6	Trabeculotomy	Pilocarpine	18	20/25	0.7	11
						GFP**	Timolol				
5	7 years	Right	38	20/30	0.7	GFP	-	12	20/60	0.7	7
		Left	37	20/80	0.4	GFP	-	8	20/20	0.4	7
6	5 years	Right	25	20/200	0.8	GFP	-	10	■	0.8	4

*: Intraocular pressure, •: Visual acuity, ••: cup-to-disc ratio, §: not available, ^^: fixes and follows, **: Guarded filtration procedure, ■: Light perception

have demonstrated the lack of perivascular nerves, confirmed immunohistochemically using antibodies to S-100 protein. (6). This is translated as altered sympathetic modulation of vascular tone, leading to progressive vascular ectasia (6). A rare cause of high intraocular pressure is the development of neovascular glaucoma, seen in association with large choroidal hemangiomas (2). We did not encounter this type of glaucoma in our patients.

Based on current knowledge of the etiopathogenesis of glaucoma in SWS, trabeculotomy is felt to be the treatment of choice when there are angle abnormalities. However, trabeculectomy should take precedence if elevated episcleral venous pressure is thought to be the cause of glaucoma, as is usually encountered in older children (7). Iwach et al (8) advocated the use of goniotomy in all patients, given the fact that trabeculectomy was associated with intraoperative choroidal expansion in 24% of their cases. However, they also found that a single goniotomy stabilized the eye for only eight months but that multiple goniotomies with medications could last up to nine years. Board and Shields (9) reported limited success after combined trabeculotomy and trabeculectomy in five eyes followed for three years. This technique proved adequate in controlling intraocular pressure in 61% of 18 patients in another series with a mean follow-up of 42 months (10). Three eyes in this study had postoperative choroidal detachments. Combined trabeculotomy-trabeculectomy has been suggested for many types of primary developmental glaucomas though the rationale for this procedure is not clear (11).

Trabeculectomy alone, with the addition of antiglaucoma drugs in some patients, has been found to be highly satisfactory in managing glaucoma that occurs “late” in patients with SWS (12, 13). Complications, however, are more common than in patients with primary open angle glaucoma or most other types. Trabeculectomy was combined with cyclocryotherapy in six patients who were followed for 3-6 years (14). The combination worked for a least six months after which timolol had to be added in two patients (14).

Mitomycin-C (MMC) has been used to enhance the effectiveness of filtering surgery in pediatric glaucoma patients, with failure and complication rates inversely related (15). Postoperative intraocular pressure control was adequate in 85% of children older than two years, as against 50% in younger ones. On the other hand, complications are more frequent in older children (15). The long-term outcome and possible side effects of MMC are still not known and we do not use this drug in our pediatric patients.

Certain potentially devastating complications including expulsive choroidal hemorrhage and massive suprachoroidal effusion may arise during or after a filtering procedure. Preoperative administration of hyperosmotic agents, strict maintenance of the anterior chamber and intraocular pressure soon after the eye is entered, tight closure of the scleral flap and sometimes prophylactic sclerostomies are suggested as precautionary measures (7, 10, 16). Trabeculotomy may be associated with hemorrhage into the anterior chamber in up to 68% of cases and bleeding may be excessive in SWS (3, 8, 11). Therefore, careful gonioscopy to identify major abnormal vessels in the angle and later, avoidance of these vessels during surgery are mandatory for the success of the procedure.

Our experience, which spans relatively long follow-up periods despite the limited number of patients, lends credence to the observation that trabeculotomy by correcting a congenital angle abnormality before age 2 years, and guarded filtration procedure, by creating a scleral opening after age 5 years, give satisfactory outcomes in terms of intraocular pressure control and stabilization of visual function. Most important, our patients enjoyed a better quality of life after a single procedure with or without antiglaucoma medication.

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